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Prenatal diagnosis of anterior duplication anomaly (dicephalus diauchenos) by ultrasonography

K. Hahmann, E. P. Issel, Christel Steinbach

Department of Obstetrics/Gynecology, Martin Luther University Halle-Wittenberg
(Director: Professor Dr. med. habil. W. Helbing)

With improved technology and increasing experience ultrasound allows early and relatively reliable diagnosis of severe fetal malformations [8, 10, 13, 18]. Double anomalies (conjoint or "Siamese" twins) are very rare [17, 19, 22, 23]. We report the case of such an anomaly (duplicitas anterior) diagnosed by means of ultrasonography.

1 Case report

Twenty-nine year old gravida 3, para 1 (in 1976 spontaneous delivery of a 2760 g boy; in 1979 a first trimester abortion); use of oral contraceptive 1976 to 1978 and following the abortion in 1979 for six months (Sequenz-Ovosiston®).
No history of significant illnesses, regular menstrual cycles. The pregnancy had an unremarkable course after the last menstrual period on 7 August, 1980. On 23 March, 1981 the patient was referred for ultrasonography in the 32nd week of pregnancy because of suspected multiple pregnancy. We used a real-time ultrasound apparatus (Sonodiagnost®, Philips) and found the fetus to be in breech position with two fetal heads in parallel position (Fig. 1) each of which had a biparietal diameter of 81 mm. There was a single chest (Fig. 2) with measurements of 86 sagital X 92 mm transverse diameter. There was a suggestion of two cardiac structures but this could not be identified definitively with the sonogram. Two vertebral columns were clearly identifiable. We assumed a fusion of the two fetuses in the area of the shoulders. The fetal movements were markedly decreased. The placenta was posteriorly located and very little amniotic fluid was present. A radiograph was of insufficient quality to add information; two heads, one trunk, two arms and two legs were identified. There was a normal fetal monitoring record. An amniocentesis for the purpose of obtaining a fetogram was initially unsuccessful. This was repeated on 15 April, 1981 in the 36th week. In the meanwhile the biparietal diameters had increased to 90 mm and the chest measurements were 94 X 102 mm. The amniocentesis yielded 5 ml of clear fluid. Subsequently, we injected 60 ml of Visotrast 370® and 10 ml Lipiodol-Ultrafluid®. The subsequent radiograph (Fig. 3) identified the intraabdominal localization of the contrast medium. Thus, one peritoneal cavity with a high diaphragm had been identified. Two days later a primary Cesarean section was carried out

Curriculum vitae

KLAUS HAHMANN *was born in 1950 and was awarded his medical diploma from the Martin Luther University in Halle-Wittenberg (German Democratic Republic) in 1976. He is now on the staff of the Women's Hospital in Halle. His area of interest is ultrasound diagnosis in obstetrics and gynecology.*

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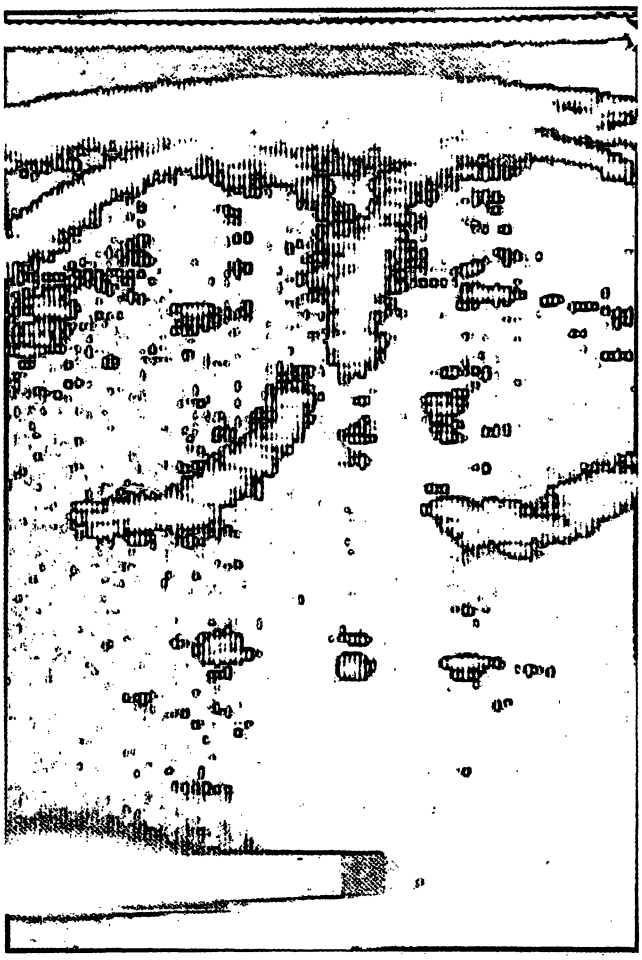

Fig. 1. Sonogram of both fetal skulls in parallel position with breech presentation.

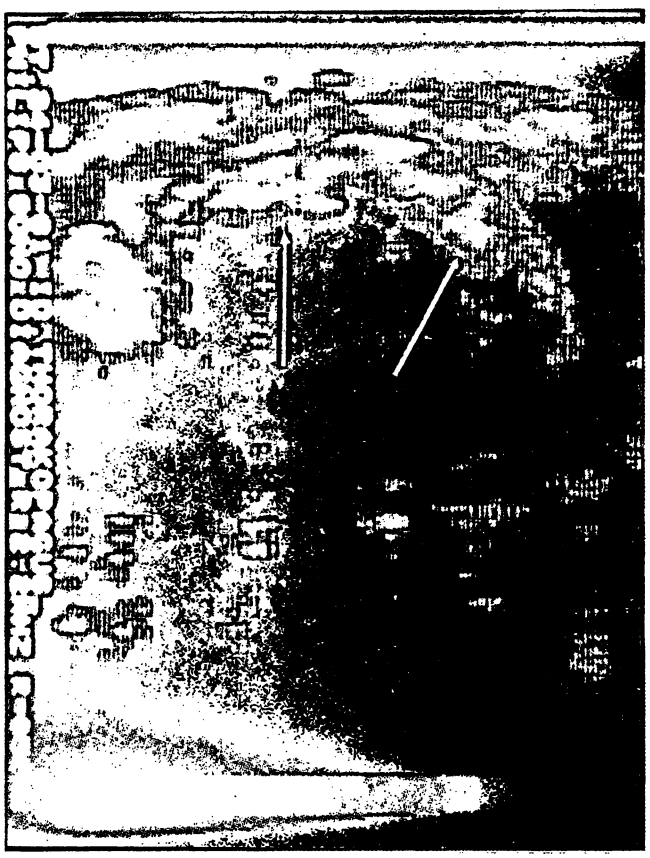

Fig. 2. Joint thoracic cavity of the two merged fetuses. The arrows indicate the two vertebral columns.

Fig. 5. Remnent of a scapula on the back. The two vertebral columns are clearly visable.

Fig. 3. Radiogram in prone position following an attempted amniofetography. The contrast medium is in the common peritoneal cavity. The arrows indicate the course of the two fetal vertebral columns.

Fig. 4. Conjoint twins (duplicitas anterior).

Fig. 6. Stump like remnent of leg in the buttock area.

after spontaneous onset of labor. The conjoint fetuses were delivered without difficulty. The mother had a unremarkable postpartum course.
The total weight of the two fetuses (Fig. 4) was 3210 grams, length each 43 cm, head circumference each 32 cm. The APGAR score at 1, 5, and 10 minutes were 2, 3 and 4 respectively. Each infant showed different respiratory movements and both cried briefly. After a course of 12 hours of increasing respiratory insufficiency both died at the same time. Thus we were able to verify the antepartum diagnosis of an anterior duplication anomaly with the absence of one arm and leg of each fetus. There were remnents of a scapula on the back (Fig. 5). The two vertebral columns were clearly visible; they merged in the lumbar area. In the buttock area, dorsally there was a stump-like leg remnant (Fig. 6). An autopsy identified the two separate hearts in a common thoracic cavity. There was a large abdominal cavity with a large liver and two stomachs and a double esophagus. There was one pair of kidneys. The infants were both girls with separate vaginae and a common uterus with one pair of adnexae.

2 Discussion

Duplication anomalies according to SCHWALBE [20] exist when there is at least a partial duplication of the trunk axis. **They are always monozygotic twins of identical sex.** The incidence is being cited as between 1:6000 to 1:500,000 [6, 12, 15, 21]. They are more commonly girls with 68% [1].
HELLMANN and PRITCHARD [7] divide the symmetrical anomalies into three groups:

1. Incomplete duplication in the upper or lower part of the body (diprosopus, dipagus).
2. Twins connected at the upper or lower part of the body (craniopagus, ischiopagus, pygopagus).
3. Twins connected at the trunk (thoracopagus, dicephalus).

The prenatal diagnosis of duplication anomalies was an exception until now and usually became apparent only because of a complication in the course of labor and delivery [3, 4, 9]. Because this results frequently in complications for mother or children the early diagnosis of duplication anomalies is clinically important.
Only a few authors have published cases of duplication anomalies diagnosed with ultrasound [2, 5, 14, 24]. All these cases were thoracopagus. We have no knowledge of an anterior duplication anomaly primarly diagnosed by a B-scan ultrasound during pregnancy. The establishment of the diagnosis causes problems for both the parents and the obstetrician. The time for delivery should be selected in consideration of the severity of the anomaly and the chance for survival of the infants by possible surgical separation. As have other authors [11, 16] we recommend for term pregnancies a Cesarean delivery.
We concur with the opinion of HANSMANN [5] that ultrasonography is currently the method of choice for the exclusion or demonstration of a fetal anomaly. We too have attempted to confirm the diagnosis with x-ray examinations. However, we had to accept that the interpretation of the pelvic x-ray poses great difficulties and that **ultimately the ultrasonography was the most accurate method.** If the diagnosis is made very early, before the 24th week of gestation, a termination of pregnancy might be considered.

Summary

A 29 year old gravida 3, para 1 was referred in the 32nd pregnancy week for ultrasonography when a twin pregnancy was suspected.
We found a fetal duplication with two heads in parallel position, biparietal diameter 81 mm (Fig. 1), a joint thoracic cavity measuring 86 X 92 mm (Fig. 2), reduced amniotic liquid and decreased spontaneous movements.
The pelvic radiogram confirmed our suspension of an anterior duplication anomaly and showed only two arms and two legs for both fetuses but brought no additional information. The amniofetography failed. The contrast media ended up in the joint peritoneal cavity of the fetuses (Fig. 3).
Delivery was in the 36th week by Cesarean section. The weight of the conjoint twins (Fig. 4) was 3210 grams, length 43 cm, and head circumference of each was 32 cm. Both fetuses showed independed respiratory movements and both briefly cried. Death occurred after 12 hours. Fig. 5 shows remanents of a scapula and Fig. 6 demonstrates a stump like leg remanent.

Our prenatal diagnosis of an anterior duplication was confirmed by the autopsy. After a prenatal diagnosis of a duplication anomaly we recommend Cesarean section in term pregnancies. Considerations when deciding on the management of the pregnancy should include survival chances following possible surgical separation. If the diagnosis is made before the 24th week termination of the pregnancy might be considered.

Keywords: Conjoint twinning, duplicitas anterior, fetal anomaly, prenatal diagnosis, ultrasound.

Zusammenfassung

Präpartale Diagnose einer Duplicitas anterior (Dicephalus diauchenos) mittels Ultrasonographie
Eine 29jährige III.-Gravida, I.-Para wurde mit 32 Schwangerschaftswochen zur Ultraschalldiagnostik mit dem Verdacht auf Zwillinge überwiesen.
Wir fanden eine doppelte Fruchtanlage mit 2 Köpfen in Parallelstellung (biparietale Durchmesser je 8,1 cm, Abb. 1), einem gemeinsamen Thorax (8,6 · 9,2 cm, Abb. 2), wenig Fruchtwasser und wenig Spontanmotorik.
Die Röntgenübersichtsaufnahme bestätigte unseren Verdacht auf Duplicitas anterior, zeigte nur 2 Arme und 2 Beine insgesamt für beide Feten, brachte sonst aber keine zusätzlichen Informationen.
Die Amniofetographie mißlang. Die Kontrastmittel gelangten in die gemeinsame Bauchhöhle der Feten (Abb. 3).
Die Entbindung erfolgte in der 36. Schwangerschaftswoche mittels Sectio caesarea.
Das Gewicht der verschmolzenen (bzw. nicht getrennten) Kinder (Abb. 4) betrug 3210 g, die Länge je 43 cm und die Kopfumfänge waren je 32 cm.
Beide Früchte zeigten unabhängig voneinander Atembewegungen und haben kurzzeitig geschrien. Der Tod trat nach 12 Stunden ein.
Die Abb. 5 zeigt Reste einer Scapulaanlage, die Abb. 6 eine stummelartige Beinanlage.
Unsere antenatal gestellte Diagnose einer Duplicitas anterior wurde durch die Obduktion bestätigt.
Bei antenataler Diagnostik einer Doppelbildung empfehlen wir bei ausgetragener Schwangerschaft die Schnittentbindung. Bei Beendigung der Schwangerschaft sollten die Überlebenschancen der Früchte nach eventueller chirurgischer Trennung in die Überlegungen einbezogen werden. Bei Diagnosestellung vor der 24. Schwangerschaftswoche kann man die Interruptio in Erwägung ziehen.

Schlüsselwörter: Antenatale Diagnostik, Duplicitas anterior, Mißbildung, Ultraschall.

Résumé

Diagnose prénatale d'une duplication antérieure (dicephalus diauchenos) par ultrasonographie.
Une femme agée de 29 ans, gravide III, para-I, a été soumise durant la 32 semaine de grossesse à un examen aux ultra-sons suite à une suspicion de jumeaux. Nous avons trouvé une duplication fétale avec 2 têtes en position parallèle (diamètre biparietal 8,1 cm, fig. 1) une cavité thorqcique commune (8,6 × 9,2 cm, fig. 2) peu de liquide amniotique et une diminution des mouvements spontanés. Une radiographie pelvienne confirma notre suspicion d'une duplication antérieure et montre seulement 2 bras et 2 jambes pour les 2 fétus, mais n'apporta pas d'information supplémentaire. L'amniofétographie n'a pas réussi. Le produit de contrast pénétra dans la cavité péritonéale commune des fétus (fig. 3). On a procédé à l'accouchement par césarienne dans la 36 semaine de la grossesse.
Le poids des jumeaux attachés (fig. 4) était de 3210 g, longueur de chacun 43 cm et le périmètre cranien était de 32 cm dans les deux cas. Les deux fétus montraient des mouvements respiratoires indépendants et ils ont tous les deux crié brièvement. Ils sont morts après 12 heures.
La fig. 5 montre des restes d'une homoplate et la fig. 6 montre un moignon de jambe.
Notre diagnose prénatale d'une duplication antérieure fut confirmée par l'autopsie.
Après le diagnostic prénatale d'une duplication nous recommendons l'accouchement par césarienne, a terme; avec la possibilité d'envisager une séparation chirurgicale qui pourrait présenter une survie. Si la diagnose est posée avant la 24 semaine de grossesse on peut envisager un avortement.

Mots-clés: Anomalie fétale, duplication antérieur, diagnostic prénatal, examen aux ultra-sons.

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Dr. med. K. Hahmann
Dr. sc. med. E.-P. Issel
Dr. med. Christel Steinbach
Universitäts-Frauenklinik
DDR-4020 Halle (Saale)
Leninallee 24